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Telemedicine in the Diagnosis and Management of Congenital Heart Disease

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1. Introduction

Several telemedicine applications have become well established in paediatric cardiology but there is potential for improvement in and expansion of existing programmes and opportunities for new applications. This chapter shall discuss why telemedicine has an important role to play in paediatric cardiology, examine current applications from fetus to adult, including the evidence base and suggest what future steps may be taken to promote the use of telemedicine in paediatric cardiology. We shall also consider what equipment and personnel are necessary for successful tele-cardiology. However, before examining the role of telemedicine in paediatric cardiology, it is first important to understand what congenital heart disease is and how it presents to clinicians.

1.1 What is congenital heart disease

Congenital heart disease can be defined as “a structural abnormality of the heart or intrathoracic great vessels that is actually or potentially of functional significance.” (Mitchell et al., 1971) The generally accepted incidence of congenital heart disease is between eight and nine per 1,000 live-born babies and this has remained consistent over several decades. (Ferencz and Neill, 1992; Royal College of Physicians, 2009) The combined incidence of moderate and severe forms of CHD is approximately 6 per 1,000 live births. (Hoffman and Kaplan, 2002) This is perhaps a more reliable and more important statistic as it accounts for babies with structural congenital heart defects that will require treatment. CHD is the most prevalent group of major congenital defects and contributes to at least 5 – 10% of neonatal deaths. (Ferencz and Neill, 1992; Wilkinson and Cooke, 1992)

1.1.1 How congenital heart disease presents

Congenital heart disease may present with a range of symptoms and clinical findings and may present at any age. However, there is certainly a trend towards earlier and even fetal diagnosis.

Symptoms: There is a wide range of cardio-respiratory and even neurological symptoms attributable to heart disease such as cyanosis, shortness of breath, syncope, presyncope and palpitations. It is increasingly common for significant CHD to be diagnosed before the patient experiences any symptoms.
Signs: It is also common for CHD to be diagnosed following the detection of an abnormal clinical sign during a routine clinical assessment or evaluation for an unrelated problem. (Table 1.1)

Imaging: Congenital heart disease may be diagnosed during routine echocardiographic screening in babies and fetuses with risk factors for CHD.

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Table 1. How congenital heart disease presents and the potential for telemedicine to support district general hospitals within a clinical network.

1.2 Why is paediatric cardiology suited to telemedicine

The single biggest reason why paediatric cardiology is suited to telemedicine is that it is a highly specialised field of medicine with a small cadre of professionals. Care is concentrated in a few regional centres. Therefore, large distances separate the patient and non-specialist from expert advice. However CHD presents, it is unlikely that a non-specialist will be confident in making a definitive diagnosis, counselling parents or instituting specific treatment without verification from a paediatric cardiologist. Fortunately, all forms of CHD can be diagnosed by a specialist using a combination of clinical examination, including stethoscopy, ECG, Chest X-Ray and echocardiographic images i.e. on the analysis of data. These data are suitable for digitisation and electronic transmission and so used in telemedicine. (Dowie et al., 2009) Babies with significant CHD may present to a hospital with on-site paediatric cardiology but considering only 13 hospitals in England and Wales have this facility compared with the 184 DGHs providing acute paediatric care, most will present to post-natal wards, neonatal units and emergency departments without on-site paediatric cardiology expertise. (Workforce Review Team, 2009)

Telemedicine can potentially support paediatricians faced with the predicament of a sick, cyanosed newborn baby with a suspected duct dependent CHD. Duct dependent lesions are a subgroup of severe CHD in which the circulation to either the lungs or body is dependent upon patency of the ductus arteriosus. This usually closes within the first 72 hours of life. In babies with duct dependent CHD, closure of the ductus arteriosus heralds the onset of severe symptoms. If not treated urgently duct dependent CHD is invariably fatal. The main differential diagnosis in this life-threatening scenario is persistent pulmonary hypertension of the newborn. The treatment of the two conditions is very different. Tele-
echocardiography interpreted by a paediatric cardiologist can offer timely diagnosis, institution of appropriate therapy and expedite or avoid transfer to a paediatric cardiology centre. (Finley et al., 1997; Casey, 1999; Mulholland et al., 1999) Antenatal diagnosis of CHD is chiefly reliant upon an abnormality being suspected by a sonographer at a DGH, with very limited experience and knowledge of CHD, during routine anomaly scanning. Telemedicine may offer support to these radiographers. Telemedicine may also facilitate a rationalisation of paediatric cardiology outreach clinics.

Paediatric cardiologists hold outreach clinics at a number of DGHs within their region, travelling on a half day or full day once a month or once every two or three months. Lower thresholds for referring asymptomatic children with clear cut innocent murmurs for “a second opinion” have swollen outreach clinics. (Dowie et al., 2009) Further pressures have been placed on service provision because of the need to comply with guidelines recommending screening of babies with various syndromes and family screening of certain inheritable conditions. Thus the paediatric cardiologist may be away from the tertiary unit on average at least once a week. Whilst an excellent service is provided at the outreach clinic, it does not seem to be efficient from the tertiary unit’s perspective. (Qureshi, 2008) It is understood that most patient’s first port of call will be their DGH. Therefore it is important to foster confidence and competence in managing children with CHD. On the other hand, patients with a clinical problem requiring a paediatric cardiology opinion that cannot wait for the next outreach clinic, need transferred to the tertiary centre for a specialist opinion.

A recent innovation has been the curriculum for paediatricians with special expertise in cardiology. Several posts have been appointed in DGHs throughout the UK. (Royal College of Paediatrics and Child Health, 2009) The role of the paediatrician with special expertise in cardiology is to unload some of the outpatient work currently being done by the tertiary centres (e.g. evaluation of children with asymptomatic murmurs or chest pain), to be the link person for children with CHD followed up at that DGH and to be competent at performing 2-D echocardiography. These paediatricians will need to be supported by the relevant tertiary centre. One of the key methods of providing this support could be via telemedicine links.

Congenital cardiac surgery for children for England is currently provided across 11 centres. Each tertiary centre serves several DGHs staffed by paediatricians. A tertiary congenital cardiology service is provided by cardiac surgeons, specialising in congenital cardiac surgery, and paediatric cardiologists. The role of telemedicine in diagnosing and excluding CHD has been recognised at a national level in the UK. Various models of care have been debated leading to a number of recommendations including the establishment of telemedicine links between DGHs and the tertiary unit in order to have echocardiograms interpreted by paediatric cardiologists at all hours, all the year round. (Qureshi, 2008) A proposed restructuring of paediatric cardiology services in the UK would reduce the number of paediatric cardiology surgical centres from eleven to six. (National Health Service, Specialised Services, 2011) Several tertiary care centres will therefore need to significantly reorganise their working practices and become part of a large network of hospitals across a wide area (Figure 1). This development, which is likely to be adopted in other countries, is proposed in order to further centralise expertise and hopefully improve outcomes for patients. However, this may disadvantage paediatric cardiologists working in non-cardiac centres and further isolate paediatricians in DGHs. The shared care of patients with CHD, between tertiary non-surgical cardiac centres and the quaternary surgical
centres, will necessitate excellent communication and case conferences. CHD case conferences are based around imaging data. It is easy to see that reliable telemedicine links between the various hospitals within this network will be essential.

![Fig. 1. The complex web of a managed clinical network](image)

In summary, congenital heart disease is potentially lethal. Its diagnosis is reliant upon interpretation of audio and visual data by a small number of experts, concentrated in a few tertiary units. Telemedicine seems to be the ideal tool to provide support to the non-specialist thereby promoting access to specialist advice and improving patient care.

1.3 History of telecardiology
Theoretically, telemedicine has been in existence since the inception of the telephone. It is a commonly held belief that Alexander Graham Bell was working on ideas to benefit the hearing-impaired when he invented the telephone in 1875. (Boettinger, 1977) In 1906, Einthoven presented his first attempts at transmitting ECGs via telephone lines. (Einthoven, 1906) The modern cardiac ambulance still utilises this application to decide upon the emergency treatment of acute myocardial infarction. In 1910 descriptions of how auscultatory sounds from a stethoscope could be amplified and similarly transmitted appeared. Apart from remote evaluation of ECGs, further developments in tele-cardiology did not materialise as the technology was not available and the clinical need not established. However, there was evidently some public interest (Figure 2).

In the 1980’s telemedicine grew steadily with developments in transmission technologies such as T-1 and ISDN and the fact that there was “something worth showing.” Developments in medical imaging during the 1980’s (e.g. echocardiography) increased diagnostic reliance on investigations whose interpretation requires an expert, usually located in a specialist centre. The first report of tele-echocardiography appeared in the 1980’s but telecardiology did not really take off in a big way until the mid 1990’s with the availability of cheaper, reliable ISDN lines and commercially available, affordable
videoconferencing codecs. (Finley et al., 1989) Telecardiology has since branched out from tele-echocardiography to other applications. In the noughties tele-homecare and remote monitoring have become the key growth areas in tele-cardiology. Research and published articles have appeared alongside each application and helped drive the field forward.

Fig. 2. Cover of Radio News magazine from April 1924 prophesising the potential role of telemedicine.

1.4 Overview of evidence available in paediatric tele-cardiology
There is a substantial body of research to support the feasibility of many applications of telecardiology in both paediatric and adult populations. There is much less good quality evidence demonstrating its clinical utility and cost-effectiveness. Most of the evidence is from small or uncontrolled studies. This is particularly difficult in paediatric cardiology because the numbers of patients involved are necessarily smaller than adult studies. However, there are larger scale randomised control trials in tele-homecare. There is now robust evidence advocating a role for telemonitoring and structured telephone support for patients with chronic heart failure. This will be discussed in more detail later in this chapter. There is also evidence suggesting that remote monitoring of pacemakers and implantable cardiac defibrillators is reliable, safe, that it may improve patient satisfaction and reduce costs. Large randomised control trials are ongoing which should hopefully clarify the role of remote monitoring in this patient group. (Burri and Senouf, 2009)

1.5 What is needed to carry out telecardiology?
The most important point to consider is: what is required from the proposed service? This should dictate the format, equipment and personnel involved. Do not start up a telecardiology service just because a particular piece of equipment comes into your possession.
1.5.1 Hardware
Videoconferencing equipment: There is not much difference from other areas of telemedicine in terms of the equipment needed. A videoconferencing codec is required at both sending and receiving ends. PC based codecs with an attached webcam were formerly favoured as they were much cheaper than purpose built, commercially available videoconferencing machines. However, through the noughties competition between the various videoconferencing companies led to significant improvements in protocols and software resulting in vastly improved videoconferencing codecs at much more affordable prices. Truly mobile videoconferencing units are now commercially available in the form of high spec webcams. This latest development is promising but has not been evaluated yet in telecardiology. Mobile phone technology is capable of facilitating many store and forward telemedicine applications e.g. simple physiological parameters and even chest-xray and ECGs. However, 3G connections are not routinely capable of facilitating medical quality videoconferencing. Mobile phones screens are also too small to conduct a video-consultation which includes a visual examination of the patient. Whatever videoconferencing equipment is employed there should be the facility to record consultations, especially those involving diagnostic imaging.
Cardiology equipment: This depends on the application. For tele-echocardiography, a standard cardiac ultrasound machine should have the requisite outputs (VGA, BNC or Phono) which will permit transmission of the echocardiographic image data to the codec. Tele-stethoscopy and tele-homecare may require more specific equipment which shall be detailed later in the chapter.

1.5.2 Network
Telecardiology grew particularly with the availability of ISDN lines which were much cheaper than what was previously available. However, in comparison with internet transmission, ISDN lines are very expensive. The majority of tele-echocardiography research has been conducted using ISDN lines and it is generally agreed that 384Kbps (ISDN6) is a satisfactory bandwidth. (Houston et al., 1999) Although no research has been conducted on this point, it is our experience that transmission of grey scale images such as cardiac angiograms requires a higher bandwidth, in the region of 1MBps. Many hospitals are part of healthcare organisations with wider computer networks affording large bandwidths for internet based videoconferencing. Thus, echocardiographic and other diagnostic images maybe transmitted at bandwidths close to 2MB. Telemedicine equipment maybe plugged into any network point in the hospital. This seems ideal for inter and intra-hospital telecardiology. However, it is still our experience that dedicated ISDN lines are the most reliable network. This advantage must be weighed against the cheaper, more flexible and higher bandwidth internet alternative. Undoubtedly the current is in favour of internet transmission.
Tele-homecare is much more fluid than inter-hospital telecardiology. Because each link is dedicated to one patient, the cost per patient is much greater than for a dedicated inter-hospital link. For home videoconferencing, ISDN links can be utilised and have the advantage of a guaranteed bandwidth but have to be installed for each home and are expensive. Whereas home broadband is already installed in many homes and in some developed countries up to two-thirds of households have internet connections with more having the availability. Home broadband connections have improved considerably in the past 10 years with larger bandwidths routinely available. Although home broadband suffers
from the problem of “contention” there are methods of “tagging” a router’s packages to give them priority traffic thereby protecting the bandwidth and videoconference quality. Therefore, home broadband is now the preferred network for conducting hospital – home videoconferences. (Horrigan, 2009)

1.5.3 Personnel
Again the question of what your telemedicine service is hoping to achieve dictates the personnel that should be involved. Inter-hospital tele-echocardiography: Two sets of personnel are required. At the sending end, a health professional is required to relay the clinical information to the receiving end. This should be the most senior doctor available. At the receiving end, a senior paediatric cardiologist is required to interpret the clinical history and transmitted images and formulate a management plan. This should preferably be a consultant but a senior trainee may also be permissible. A health professional is also required to acquire echocardiographic images. Published literature is divided into studies where an adult cardiac sonographer has performed the echocardiogram and those where a paediatrician has acquired the images. (Finley et al., 1997; Mulholland et al., 1999; Tsilimigaki et al., 2001; Sable et al., 2002; Widmer et al., 2003) There are arguments in favour of either method. One of the major obstacles to performing tele-echocardiography is the lack of personnel who can use a cardiac ultrasound machine. It requires quite a lot of practice to become familiar with the controls not to mention how and where to position the probe in order to acquire images. An adult cardiac sonographer may not perform echocardiograms on neonates but they are very proficient at using a cardiac ultrasound machine and the standard probe positions are common to both paediatric and adult echocardiography. Therefore, little guidance is required from the paediatric cardiologist. This method is well suited to a store and forward protocol. However, many DGHs do not have out-of-hours cardiac sonographer cover which could limit tele-echocardiography to a nine to five, Monday to Friday service which acute medicine most certainly is not.

Alternatively, the attending paediatrician / neonatologist could receive some training in performing echocardiography and acquire the relevant images for the tele-consultation. This requires more patience and input from the paediatric cardiologist in terms of directing the paediatrician which way to position the probe and pointing out the relevant cardiac structures as they come into view. Therefore, real-time telemedicine is much better suited to this method. With the paediatrician performing the echocardiogram, there is now the possibility of a 24 hours a day, seven days a week telemedicine service which is obviously preferable. It also provides opportunities to educate and re-enforce good practice. However, it is important that scanning skills are refreshed. For these reasons, we prefer the paediatrician to be actively involved in performing the echocardiogram as opposed to adult cardiac sonographers but acknowledge that there are advantages to the latter.

Tele-stethoscopy: The drive behind tele-stethoscopy is to produce remote paediatric cardiology out-patient clinics. These have been successfully set-up in neurology, ENT and Dermatology to name but a few. (Patterson, 2001)(Levin and Warshaw, 2009)(Dorrian et al., 2009) A consultant paediatric cardiologist is required at the receiving end as he/ she has to ultimately take responsibility for the consultation. At the sending end, a nurse could be trained to take a history and perform an ECG (if necessary), then direct the camera for a visual examination followed by placing the tele-stethoscope in the requisite positions for
auscultation. This telemedicine application does not necessarily require a doctor at the transmitting end. However, this would be a rare and excellent opportunity for continuing professional development.

Tele-homecare: In the home, only the parent/primary carer along with the patient are required. However, it is useful for primary care staff such as the health visitor or community children’s nurse to be occasionally present. This allows the non-specialist to see what the paediatric cardiologist focuses on during a consultation and fosters good working relations. It is important that tele-homecare delivered by the tertiary unit does not undermine the primary care team. In the hospital, the tele-consultation should be taken by a health professional experienced in paediatric cardiology and familiar with each patient’s diagnosis and history. This may be a paediatric cardiology trainee or an experienced nurse such as a cardiac liaison nurse. It is important that the consultant paediatric cardiologist is kept informed of developments with his/her patients and is supportive of the staff involved in the tele-homecare programme.

1.6 Economic evaluation
The area most neglected in telecardiology research is economic analysis. There have been even fewer economic analyses of telemedicine applications in paediatric cardiology. The Royal Brompton Hospital, London, have published a series of articles whose main interest has been economic evaluation of a paediatric telecardiology service. This service encompasses real-time and store and forward telemedicine for fetal tele-echocardiography, neonatal tele-echocardiography and paediatric cardiology outpatient services. Using Bootstrapping analysis, costs were calculated for the first six months after the initial telemedicine consultation. Across all four hospitals within the telemedicine network, the mean cost of implementing and running the telemedicine service is higher at each time interval than conventional referral. However, owing to the large standard deviation, these differences are not statistically significant. This analysis of the full range of paediatric tele-cardiology services is valuable. However, the amalgamation of services evaluated, the different populations involved (fetuses, neonates and children) and the different telemedicine formats employed make it difficult to elicit generalisable conclusions. For any paediatric cardiology centre considering embarking on a telemedicine programme it is perhaps more helpful to individually assess the separate components of a tele-cardiology service. To this end, the Brompton group describe an economic evaluation of their experience of a fetal tele-cardiology service. The authors conclude that the telemedicine assessment was more costly than a face-to-face examination in London (p < 0.001). However, using bootstrapped analysis, the telemedicine service becomes cost neutral at 14 days and six months. This study allows the reader to make more definite conclusions as to what the costs and savings of providing a similar fetal tele-cardiology service in their own region.(Dowie et al., 2007; Dowie et al., 2008; Dowie et al., 2009) The only other economic evaluation relating to paediatric cardiology reported in the literature is a cost minimisation study of a paediatric cardiology telemedicine network in Quebec, Canada. Whilst the authors describe this study as a cost-effectiveness analysis, no cost per health outcome is presented. This study suggests that telemedicine care is more expensive than conventional care.(Sicotte et al., 2004)
However enthusiastic telemedicine initiatives begin many are not sustained. One of the reasons for the short term nature of telemedicine projects is a lack of long-term funding which is partly due to sparse quantitative evidence including cost analyses.(Smith et al.,
2007) Previous studies reporting feasibility and diagnostic accuracy have included cost analysis as a secondary outcome. (Mulholland et al., 1999; Sable et al., 1999) However, these cost analyses were methodologically flawed, being cost-minimisation studies based on small scale, start-up programmes.

Another criticism of cost analyses involving telemedicine projects is that they are based upon start up programs where the initial burst of interest and activity decreases the unit cost of a teleconsultation. (Roine et al., 2001) Telemedicine cost analyses have been criticised for being small scale, short term pragmatic evaluations. In a systematic review of cost effectiveness studies in telemedicine, Whitten et al concluded that there is no good evidence that telemedicine is cost effective. (Whitten and Cook, 1999) In fact there have been very few cost-effectiveness studies because by definition this requires measuring a cost per unit of health outcome e.g. QALY. Most cost analysis to date have been cost minimisation studies which are out of favour and not likely to persuade health authorities to fund telemedicine services.

Payment for telemedicine consultations remains problematic. In our healthcare model (UK National Health Service) there is no mechanism in place for the regional unit to receive payment for each remote consultation. However, it maybe possible to build telemedicine services into the job plan of specialists in recognition of time undertaken in remote consultations. This is not a satisfactory situation. Other healthcare organisations, for example in the USA, have the potential to charge the referring institution a fee for each telemedicine consultation. Up to 12 states in the USA now mandate that health care plans cover telemedicine.

In summary, despite the presence of many paediatric cardiology telemedicine programmes, there is really very little evidence supporting its use from an economic perspective. This seems paradoxical as one of the founding hopes of telemedicine was that it would be cost-effective. In reality, the main problem with economic evaluation of paediatric telecardiology is a lack of well conducted studies!

2. Telemedicine applied to specific cardiology situations

2.1 Telemedicine in fetal cardiology

There are a number of features of fetal cardiology that appear to make telemedicine an attractive proposition:

2.1.1 The clinical imperative

1. Fetal cardiology is a highly centralized subspeciality with even fewer specialists than paediatric cardiology.
2. Patients often travel large distances (up to 160 km in Northern Ireland) to attend a fetal cardiology clinic. The distance between home and the specialist centre is even greater in larger countries. In Quebec, 34% of mothers referred to a fetal cardiology service travel more than 100km. (Cloutier and Finley, 2004)
3. The summary of fetal cardiology literature demonstrates that timely, antenatal diagnosis of CHD is important for improving outcomes for fetus and family by facilitating: parental counselling, elective delivery at an appropriate centre, parental choice in terms of termination and permitting fetal intervention in terms of pharmacotherapy for dysrhythmias and catheter intervention for severe complex lesions. (Bonnet et al., 1999; Allan, 2000; Tworetzky et al., 2001; Gardiner, 2008)
4. The vast majority of fetuses with CHD are born from pregnancies without a recognisable antenatal risk factor. Obviously, formal evaluation by a fetal cardiologist cannot be offered to every single pregnancy. Therefore, in order to maximise antenatal detection of CHD, greater proficiency, from the non-specialist, is required at confirming normality and identifying pathology.

In the UK, the most practicable method of promoting antenatal detection of CHD is by improving cardiac evaluation during the routine anomaly scan. Previous studies have demonstrated the benefit and relative ease with which more detailed imaging of the heart may be incorporated into routine anomaly scanning. (Stumpflen et al., 1996) However, such reports often reflect working practices immediately after the introduction of a training program. As the teaching study from Hunter et al demonstrated, there is a danger that if support from paediatric cardiology is not ongoing then the initial efficacy of training may diminish. (Hunter et al., 2000) Telemedicine has the potential to facilitate earlier diagnosis of CHD by offering increased and quicker access to specialist opinion. Telemedicine may also support sonographers’ education and training.

Fetal cardiology is predominantly a speciality of medical imaging as physical examination of the fetal cardiovascular system is obviously not possible and fetal catheter and surgical interventions are limited to a handful of centres worldwide. As is well documented, telemedicine lends itself to areas where medical imaging is crucial to diagnosis. (Casey, 1999) It may be suggested that performing fetal tele-echocardiography is much the same as tele-echocardiography in neonates but fetal echocardiography is significantly different from neonatal echocardiography:

1. The fetal heart is smaller and beats faster which may exacerbate motion artefact.
2. The fetus is surrounded by the mother so that ultrasound has to travel through more tissue impacting on image quality.
3. The fetus may move.
4. The fetus may be in a disadvantageous position e.g. back up.

Experience of telemedicine in fetal cardiology is still in its infancy. The fetal cardiology telemedicine programs that are currently active appear to have developed from centres with a major interest in telemedicine and fetal medicine. Whilst an acceptable bandwidth (384kbps) has been established for neonatal tele-echocardiography. This is not the case with fetal tele-echocardiography. Therefore, research is required in this area.

2.1.2 Evidence for fetal tele-cardiology

Despite a potential role for telemedicine in fetal cardiology, there are very few studies investigating its potential. Only one study is reported to date that investigates the feasibility and acceptability of performing remote, real-time 2-D fetal echocardiography. Sharma et al performed a study in two phases. Phase one was a laboratory study, investigating the effect of bandwidth on image quality and overall adequacy of scan. Sixty-four fetal echocardiograms were performed and randomly assigned to transmission across 128, 384 and 768 kbps and recorded on Super VHS videotape for subsequent review. Six studies were excluded due to the presence of CHD. Of the 58 real-time studies interpreted, 15 / 58 were transmitted across 128 kbps, 21 across 384 kbps and 22 across 768 kbps. The image quality was felt to be significantly poorer at 128 kbps. At 128 kbps, the mean Likert score was 1.1/5 compared with 3.1 / 5 at 384 kbps (p < 0.01) and 3.4 / 5 at 768 kbps (p < 0.01). There was no significant difference in perceived image quality in transmission across 384 kbps compared with 768 kbps (p = 0.08). Similarly, there was no significant difference in the
Sharma et al conclude that 384 kbps is an adequate bandwidth for the identification of normal heart structures. In the second phase of the study, 34 fetal echocardiograms were successfully transmitted and interpreted in real-time. In four cases (11%) technical problems prevented transmission. The fetal echocardiogram was performed by a fetal cardiac sonographer or a paediatric cardiology fellow with experience in fetal echocardiography. This was compared with a video recording of the study made at the transmitting site. Mothers were followed up with a face-to-face fetal echocardiogram if a concern was identified on the transmitted fetal echocardiogram. In order to evaluate patient satisfaction, a questionnaire was completed by all 34 expectant mothers and a control group of 195 patients referred from peripheral hospitals to the regional fetal cardiology clinic. There was no significant difference in the completeness of each study between the tele-fetal echocardiogram and the recorded fetal echocardiogram. CHD was identified correctly in two transmitted fetal echocardiograms with a third case of CHD being suspected but only confirmed on direct fetal echocardiography.

The satisfaction survey results were very positive. In all nine questions, the mean patient satisfaction score indicated high levels of satisfaction with the telemedicine consultation. In eight questions there was no significant difference in patient satisfaction between the telemedicine and control groups. In addition, only 6% of patients stated that they felt uncomfortable talking to the doctor across the tele-link and only 3% felt it was difficult to ask questions.

In this important study, the authors conclude that transmission of fetal echocardiograms across 384 kbps is feasible and adequate for the interpretation of screening fetal echocardiograms. Community acceptance of telemedicine screening and counselling is not adversely affected by a lack of direct personal contact with the specialist. Whilst this study is valuable there are some limitations. The majority of transmitted fetal echocardiograms were not followed up with a face-to-face fetal echocardiogram which is the gold standard. The proportion of fetuses with CHD was small in this cohort. Therefore, the diagnostic accuracy of the telemedicine process was not adequately assessed. The fetal echocardiograms were performed by an operator already skilled in fetal echocardiography. This may falsely enhance the adequacy and quality of the fetal echocardiogram being transmitted. It is suggested that most difficulties in interpreting echocardiograms transmitted via a tele-link are related to image acquisition rather than transmission (Cloutier, 2000). Eleven per cent of studies were terminated due to technical difficulties. A routine clinical service would be significantly limited if 11% of consultations were aborted due to technical difficulties.

There are also reports of telemedicine facilitating 3-D fetal echocardiography by spatio-temporal image correlation (STIC) and datasets transmitted across an ADSL link. STIC is a relatively new approach for clinical assessment of the fetal heart. It is an easy technique for acquiring data from the fetal heart (a single, automatic volume sweep) and produces a 4D cine sequence. A group from Chile conducted a similar study STIC transmitted across an ADSL connection with bandwidths of 300 and 600 kbps. In total fifty scans were transmitted from two peripheral hospitals to the regional centre. On average complete fetal echocardiograms could be interpreted in 92%.(Vinals et al., 2005) However, these isolated studies have not been followed up in the literature.

In a study conducted in our own department we evaluated fetal echocardiography, performed by sonographers with little training in fetal cardiology, but guided by a fetal
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cardiologist and transmitted in real-time across ISDN 6 (Figure 3). The remote consultation concluded with a videoconference in which the fetal cardiologist explained the diagnosis and answered any questions from the prospective parents. This fetal-telecardiology consultation was compared with the subsequent “face-to-face” fetal cardiology consultation at the regional centre. Sixty-eight fetal tele-echocardiograms were performed and followed-up. The diagnostic accuracy was very high (κ statistic = 0.89, sensitivity = 0.92, specificity = 0.98, ppv = 0.92, npv = 0.98) (Figure 4). Sonographers were very positive regarding their involvement in the study and reported significantly improved levels of confidence in performing fetal echocardiography. A significant positive feature of real-time fetal tele-echocardiography is the continued support provided to radiographers performing fetal anomaly scans in DGHs. Patients were very positive about the telemedicine process and were equally satisfied with the remote consultation as the face-to-face consultation. Patients were equally satisfied with each consultation but consistently replied that they would prefer to have the fetal cardiology consultation performed at the local hospital facilitated by a tele-link rather than a face-to-face consultation at the regional centre. (McCrossan, 2009)

Fig. 3. What is seen by the paediatric cardiologist a) Live guidance of fetal echo, b) Counselling parents

During the study it was noted that the received fetal echocardiographic images were of poorer quality following an upgrade in the ultrasound scanner. This did not appear to affect diagnostic accuracy. With the superior imaging, this deterioration in received picture quality was probably due to excess data being transmitted down the 384kbps connection with resulting package loss and pixellation. This problem would likely be reduced if a larger bandwidth was employed. Extensions to inter-hospital networks make this a realistic option. Alternatively SDSL links (2MBps) can be installed at much cheaper rates than corresponding ISDN bandwidths.

The introduction of a remote fetal cardiology service would have attendant costs which are not offset by savings in inter-hospital transfers. However, the possible establishment of a “one-stop-shop” for “high-risk” pregnancies in which the routine fetal anomaly scan is followed by a remote fetal echocardiogram confidently identifying all relevant cardiac

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structures (55% in this cohort) could potentially be cost neutral, relieve pressure on the regional unit and present time and cost savings to the patient. Ultimately the aim of future work should be aimed towards the establishment of a regional fetal tele-echocardiography service.

Fig. 4. Example of severe congenital heart disease accurately diagnosed via tele-link in this study.

2.2 Tele-echocardiography in paediatrics
The main application of telemedicine in paediatric cardiology to date has been remote diagnosis of congenital heart disease by the transmission of echocardiograms from DGH to specialist. There are many articles in the literature confirming the effectiveness of this telemedicine application.(Finley et al., 1997; Mulholland et al., 1999; Sable, 2003; Widmer et al., 2003; Grant et al., 2010)(Tsilimigaki et al., 2001)

2.2.1 Clinical imperative
1. It can be challenging to confidently rule out CHD in the differential diagnosis of a critically ill newborn baby. In these cases, paediatricians often find it difficult to confidently exclude CHD.
2. Many such babies present at DGHs with no paediatric cardiology support on site.
3. The treatment of CHD can be very different from that of other life-threatening conditions. Therefore making the correct diagnosis can be life-saving.
4. Sick newborn babies without CHD maybe cared for very well at the DGH and should be spared a potentially hazardous medical transfer.

2.2.2 Evidence for neonatal tele-echocardiography
The literature demonstrates that neonatal tele-echocardiography is feasible and reliable. High diagnostic accuracy is reproducible with live guidance by a paediatric
cardiologist. (Mulholland et al., 1999; Sable et al., 1999; Widmer et al., 2003; Grant et al., 2010)

Store and forward formats have also been utilised but in this situation the paediatric cardiologist must be confident that the echocardiographer will obtain a detailed and complete echocardiographic study. If the echocardiographer is capable of performing such a study it is questionable whether further interpretation is necessary. More recent studies also demonstrate that tele-echocardiography helps avoid potentially risky transfers of sick newborn babies between hospitals. (Grant et al., 2010) Most importantly, neonatal tele-echocardiography has become accepted as an integral part of many regional paediatric cardiology services across the world.

The value of telemedicine in continuing professional development has been well documented if not fully researched. (Casey, 1999; Sable, 2002) Our experience of the educational benefit of real time transmission of echocardiograms has been very positive. Having performed echocardiograms under the guidance of a paediatric cardiologist, paediatricians report an improvement in their ability to interpret echocardiograms and have been observed to gain confidence in echocardiography. We believe that real-time, live guidance of the paediatrician by the paediatric cardiologist has advantages compared to a "store and forward" protocol, in both acquiring quality images and in providing an educational support. Often the inexperienced sonographer obtains images that they believe are not adequate. However, it is a difficulty interpreting the images that is the major contributing factor to poor image acquisition. During the videoconference, the paediatric cardiologist is able to highlight relevant structures as they appear and describe what manoeuvres are necessary to view other structures.

There is an important caveat to tele-echocardiography. In a small number of cases major CHD will be suspected following the transmitted scan but a definite diagnosis cannot be established. These cases highlight the importance of recognising the limitations of telemedicine. If there is a clinical suspicion of major CHD that cannot be confidently diagnosed or excluded, following a tele-echocardiogram, then transfer to a regional unit for hands-on assessment should not be delayed.

Ideally, we envisage a hub and spoke model for the provision of paediatric cardiology with tele-echocardiography acting as a valuable aid in supporting a paediatrician with special expertise in cardiology.

Economic analysis

As discussed previously, economic analysis of telemedicine applications is lacking and this holds true for neonatal tele-echocardiography. However, a study performed at our own institution demonstrates clearly that neonatal tele-echocardiography is cost-saving for each DGH within the telemedicine network. Although there are significant start-up costs for each hospital, these are outweighed by the savings made from avoiding expensive inter-hospital transfers as a result of the remote consultation. This study was conducted over an eight year period and suggests that telemedicine services can continue to be cost saving beyond the initial enthusiasm surrounding a new service. Northern Ireland is a small geographical area, therefore the potential saving for larger regions is even greater. (Grant et al., 2010)

One of the main barriers to increased use of telemedicine is the initial start up cost. Running costs for ISDN 6 lines are also significant. As predicted, the cost of videoconferencing codecs suitable for transmission of echocardiograms has decreased significantly over the past 10 years. More recently, the possibility of using the internet as a means of transmission has been explored with some success. This may be achieved through a healthcare organisation’s
wide area network or by installing individual internet links such as an SDSL connection (Up to 2MBps). Our experience of ADSL (in its current provision) is that the variable bandwidth is not suitable for tele-echocardiography. As comparable internet links are much less expensive than ISDN lines, switching to internet connections will only help the economic argument in favour of telemedicine.

Payment for telemedicine consultations remains problematic. In our healthcare model (UK National Health Service) there is no mechanism in place for the regional unit to receive payment for each remote consultation. However, it maybe possible to build telemedicine services into the job plan of specialists in recognition of time undertaken in remote consultations. This is not a satisfactory situation. Other healthcare organisations, for example in the USA, have the potential to charge the referring institution a fee for each telemedicine consultation. Up to 12 states in the USA now mandate that health care plans cover telemedicine. This seems a more sustainable approach to telemedicine and should be possible in any healthcare organisation as payments are routinely made between health boards for face-to-face medical consultations.(Macios, 2010) This seems a more sustainable approach to telemedicine and should be possible in any healthcare organisation as payments are routinely made between health boards for face-to-face medical consultations.

2.3 Tele-homecare in paediatric cardiology

2.3.1 Background

Paediatric cardiology has changed dramatically over the past twenty years. Improvements in diagnostic imaging and innovations in surgical technique along with intensive care, have revolutionised the prognosis for children with major congenital heart disease.(Laussen, 2001; McElhinney and Wernovsky, 2001) In particular children with complex CHD, typified by single ventricle physiology, now have the possibility of life beyond the neonatal period.(Marino, 2002) However, such patients are not “cured” but receive long-term palliation in the form of a staged surgical programme over several years.

Parents often state that the most anxious and stressful period is not around the time of diagnosis or even surgery but actually the day they take their baby home for the first time. Parents feel very reassured by the presence of nursing staff and the 24 hour availability of medical expertise which is only possible in hospital.(Holmes, 1996)

The clinical imperative

1. Patients with severe congenital heart disease require particularly careful follow-up in early infancy and it seems logical that home support / monitoring could potentially contribute to the quality of care delivered.

2. At discharge there may be ongoing clinical issues which may not require medical attention but are an added stress to parents e.g. feeding difficulties, ongoing cyanosis and inter-current viral infections. In this setting, it is understandable that parents are extremely anxious and in want of support and reassurance.

3. It is recommended that tele-home care be targeted at conditions that require close monitoring, clinical assessment and early intervention to avoid adverse events such as hospitalization or emergency visits.(Dellitrafine and Dansky, 2008)

4. Paediatric cardiology is a highly specialized and centralized field of medicine. The paediatric cardiology team who have been involved in the daily management of these patients are ideally placed to monitor the patient’s progress and deal with any problems as they arise.
2.3.2 Evidence for tele-homecare in paediatric cardiology

The group of congenital cardiac lesions that have attracted most interest over the past 15 years have been those which fall within the umbrella term - “hypoplastic left heart syndrome.” In HLHS, there is severe under development of some or all left heart structures.(Dhillon and Redington, 2002) Until relatively recently, surgical intervention was not widely available. However, it is now exceptional for babies with HLHS not to be offered surgical intervention. This usually takes the form of staged surgical palliation culminating in a total cavo-pulmonary anastomosis with a systemic right ventricle. The first stage is the Norwood procedure, performed during the early neonatal period, with the second stage occurring during mid-infancy.(Bove et al., 2004; Sano et al., 2004; Stasik et al., 2006) Although modifications to the Norwood procedure have been associated with a reduction in early mortality, there remains a significant attrition rate between the first and second stages.(Forbess, 2003) This group of patients could particularly benefit from additional monitoring and support following discharge from hospital.

A group from Wisconsin, USA, postulated that patients at risk of inter-stage mortality could be identified by a deterioration in their physiological status. A daily home programme of measuring arterial oxygen saturations and weight was combined with a protocol guiding the need for hospitalisation. There was significantly less mortality observed during the 15 month intervention period compared with the preceding 50 month control period (0% vs 16%, p = 0.039).(Ghanayem et al., 2003; Ghanayem et al., 2004; Ghanayem et al., 2006)

We conducted a randomised control trial of a home support programme facilitated by videoconferencing compared with telephone support and a control group for babies with major CHD. With the assistance of a videoconferencing company, we installed commercially available, portable videoconferencing systems in the patients’ homes and included pulse oximetry when clinically indicated. ISDN lines were initially utilised but this was switched to an ADSL connection during the latter half of the study. Regular remote consultations were conducted in both intervention groups (Figure5). The results of this study demonstrated that home support delivered by videoconferencing is feasible, reliable, sustainable and superior to telephone support in terms of reducing parental anxiety. Moreover, videoconferencing home support was associated with significantly reduced health service utilisation and may possibly be cost-saving.(Morgan et al., 2008; McCrossan, 2009) Our institution has secured government funding to incorporate videoconferencing home support into the post-discharge care for a subset of babies with major CHD.

2.3.3 Tele-homecare in adult cardiology

There is a wide array of tele-care projects and services available. In adult cardiology, most tele-monitoring services are centred around the regular transmission of vital parameters (Blood pressure, heart rate, respiratory rate and weight) from home to hospital. There are many tele-homecare systems specifically designed for this purpose. Variations in the patient’s observations outside set limits trigger a response from the hospital which may involve changing medication etc. Other services are based upon structured telephone support which is usually nurse led. Web-based patient education often forms a component of tele-health services.

There is good and increasing evidence that tele-homecare can be an effective method of health care delivery. A recent meta-analysis of all home based telehealth research indicated a moderate positive effect on health outcomes (mean weighted effect size = 0.5, p<0.01). Sub-analysis revealed particular benefits in patients with mental health illness and chronic adult heart disease but no significant effect in diabetes mellitus.(Dellifraine and Dansky,
The potential for tele-homecare in the management of chronic obstructive pulmonary disease has also been highlighted and trials are now in process (McKinstry et al., 2009; Lewis et al., 2010). Home monitoring is now established as an adjunct to the ongoing management of specific adult cardiac populations: e.g. chronic heart failure, hypertension and implantable cardiac defibrillators / pacemakers (Clark et al., 2007; Cleland et al., 2005; Heidbuchel et al., 2008; Raatikainen et al., 2008; Parati et al., 2009). This has culminated in a Cochrane review confirming that telephone support and tele-monitoring programmes for patients with congestive heart failure are effective in reducing the risk of all-cause mortality and CHF-related hospitalisations. Furthermore, such programmes improve quality of life and reduce costs (Inglis et al., 2011).

Following the accumulation of good quality evidence, government funding has been forthcoming. The EU has invested more than €650 million in funding tele-health and telecare initiatives (Celler et al., 2007). Unsurprisingly, tele-homecare projects are the main growth area in telemedicine within cardiology. In the context of current healthcare models, this is set to continue.

2.3.4 Future

There is a strong case for conducting similar research trials in other paediatric subspecialties e.g. ex-preterm infants, chronic respiratory and neurological patients. The central premise is common: complex, vulnerable patients scattered across a wide geographical area who are likely to benefit from regular, direct input from the tertiary care team. The scope of home monitoring could also be increased to include ECG monitoring for patients with dysrhythmias and patients requiring INR monitoring for warfarin. A significant proportion of a cardiac liaison nurse’s workload could be facilitated by a telemedicine home support mechanism. Such a scheme could potentially reorganise the role of the cardiac liaison nurse and provide more efficient use of resources. Advances in home broadband links in terms of...
quality, geographical coverage and cost will facilitate more ambitious tele-homecare programmes with the potential to improve patient care.

2.4 Tele-stethoscopy

2.4.1 Clinical imperative

1. Innocent murmurs are very common during childhood. Whilst definitive studies have proved elusive, the estimated prevalence in developed countries is approximately 50%. However, congenital heart disease diagnosed following the detection of a murmur after infancy is rare in comparison. Approximately 2% of patients with a murmur presenting after 12 months old have CHD (O'Rourke, 2004).

2. Paediatric cardiology outpatient clinics are over subscribed and there is limited capacity for extra clinics at a regional unit.

3. Outreach clinics are an important aspect of a regional service but take the paediatric cardiologist away from the tertiary unit. As there are only a small number of paediatric cardiologists this can be a significant consideration in terms of providing senior cover at the tertiary centre. It is possible that remote consultations could be substituted for face-to-face consultations. Alternatively telemedicine may streamline referrals. In both situations the auscultatory findings are paramount.

2.4.2 The role of auscultation in paediatric cardiology outpatients

Traditionally, clinical examination has been the primary method of assessing children referred for evaluation of an asymptomatic murmur. Prior to the advent of 2-D echocardiography, reliable, non-invasive imaging of the heart was not possible. With improvements in computer software, enabling superior 2-D imaging, colour flow mapping, Doppler measurements, and the increased availability of ultrasound scanners, comes the temptation to routinely employ echocardiography. However, there is good evidence that clinical examination alone is highly sensitive and specific.

The question is: who should perform this assessment? There are numerous studies in the literature examining the roles of the non-specialist and paediatric cardiologist in the initial evaluation of children with asymptomatic cardiac murmurs. The available literature suggests that non-specialists are better than they think at detecting congenital heart disease achieving good levels of specificity. However, they are much less confident at excluding congenital heart disease. (Rushforth and Wilson, 1992) The result is a high volume of referrals of patients with innocent murmurs. Anecdotally, this lack of confidence in distinguishing innocent from significant murmurs has increased over the past 30 years. (Noonan, 1999) The reasons for this may be due to concerns over malpractice, greater availability of sophisticated investigations and a misperception that echocardiography is necessary to rule out pathology. (Danford et al., 1993) Therefore we have a situation where there is a highly centralised service with limited capacity in which the key feature of the consultation is auscultation. Sound waves may be captured, processed and transmitted for evaluation at a distance. The key question is: can heart sounds be reliably sampled and transmitted for accurate interpretation at a remote location?

2.4.3 The evidence for tele-stethoscopy

Although remote stethoscopy was reported as early as 1973, the first significant study, appeared in the 1990s. In an elegant study, Belmont et al demonstrated that real-time tele-
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Stethoscopy was accurate and clinically useful ($\kappa > 0.6$) for detecting murmurs, distinguishing specific murmurs (innocent, pathological, vibratory, diastolic aortic, diastolic pulmonary) and diagnosing congenital heart disease ($\kappa = 0.63$). Interestingly, Belmont found that remote stethoscopy’s accuracy suffered in younger children ($\kappa = 0.29$). Consequently, Belmont et al conclude that remote stethoscopy provided accurate, dependable detection of congenital heart disease but only in children five years old and above. The feasibility and accuracy of real-time tele-stethoscopy have been supported by further studies including research at our own institution. (McConnell et al., 1999; Grant, 2006; McCrossan, 2009)

Alternatively, tele-stethoscopy maybe achieved using a store and forward protocol. Digital stethoscopes are capable of sampling and recording heart sounds which can then be e-mailed to the paediatric cardiologist along with the history and additional clinical findings. Several studies, using this technique, have been published which demonstrate similarly high levels of accuracy in differentiating innocence from pathology i.e. the need for further investigation in the form of echocardiography. (Dahl et al., 2002; Finley et al., 2006; Mahnke et al., 2008)

Although the Belmont study suggested that tele-stethoscopy was significantly less accurate in children under 5 years, more recent reports in the literature and our own experience do not support this claim. We would suggest that children three years and over are suitable for tele-stethoscopy i.e. the pre-school child with a murmur.

There are arguments in favour of both real-time and store and forward protocols in the application of tele-stethoscopy. Real-time stethoscopy requires more elaborate telemedicine equipment which has historically been bespoke PC-based systems. Such home-built systems are temperamental and difficult to replicate. However, there are now better quality, commercially available tele-stethoscopy systems which are more user friendly (Figure 6). Real-time tele-stethoscopy is more time consuming and requires greater co-ordination. However, the audio-visual contact with the patient and parent is probably beneficial in terms of reassuring parents. There is also the opportunity to repeat auscultation and perform respiratory and positional manoeuvres which may tease out the character of a murmur. Whilst this is possible with store and forward sampling, not every aspect of auscultation is performed with every consultation. The course of a clinical examination is influenced by what the clinician believes is the differential diagnosis on first hearing the murmur / heart sounds. Therefore, either a large number of samples are recorded for each patient, which must increase the number of inadequate samples, or a limited number of samples are recorded which may leave the paediatric cardiologist wishing that he/ she could have another listen. However, there is an undeniable advantage to sitting in an office, reading the history / examination, viewing the ECG / CXR and then listening to a few audio files compared with setting up and co-ordinating a remote clinic. For this reason, the future of tele-stethoscopy, as a clinical tool for out-patient referrals, is likely to rest with the store-and-forward technique.

Adult cardiology does not have the same need for tele-stethoscopy as paediatric cardiology. Ischaemic heart disease rather than structural defects comprise the bulk of the workload so murmurs do not have equivalent significance. Additionally, most DGHs have adult cardiology specialists with access to echocardiography. However, there are reports of real-time tele-stethoscopy from an Italian group which they believe should be useful in the setting of tele-homecare. Fragrasso et al employed a commercially available, purpose built videoconferencing tele-stethoscopy system. They report that cardiac and respiratory auscultation was accurate and valuable. (Fragrasso et al., 2007)
Fig. 6. (a and b) Commercially available tele-stethoscopy system from Aethra and (c) Digital stethoscope

3. Conclusions

The research presented in this thesis demonstrates that telemedicine may be applied across multiple common paediatric cardiology scenarios: from fetus, to neonate, to infant to child; and from home, to outpatient clinic. A wide range of clinical data, including visual examination, auscultatory sounds, physiological parameters and echocardiographic images, may be transmitted in real-time across telemedicine links facilitating accurate diagnosis, effective decision making and promoting access to specialist opinion. Telemedicine is an acceptable process to both health professionals and the public. It is associated with high levels of parental satisfaction. Telemedicine applications are shown to impact on patient management and reduce health service utilisation with the net effect of keeping patients and their families closer to home. Established telemedicine programmes may continue to be cost saving beyond the initial start-up phase. Internet transmission is likely to improve significantly in the future and is economically more sustainable compared with ISDN.

In this chapter I have not addressed the issue of who takes legal responsibility for the patient following the telemedicine consultation. Despite almost 20 years of experience of
telemedicine in paediatric cardiology, there are no agreed guidelines concerning professional responsibility. Currently, it is likely that all physicians involved take responsibility for their own professional conduct in relation to the patient. To date there have been no cases of medical negligence resulting from telemedicine consultations in the UK. This may conceal out of court settlements. (Avienda Telemedicine Consultancy, 2007) The main recommendation from this chapter is for clinicians (for only clinicians can make telemedicine work) to consider whether areas of their clinical practice are negatively affected by distance separating themselves from either patients or colleagues. If so telemedicine may facilitate more effective and efficient use of resources and improve patient care.

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Telemedicine is a rapidly evolving field as new technologies are implemented for example for the development of wireless sensors, quality data transmission. Using the Internet applications such as counseling, clinical consultation support and home care monitoring and management are more and more realized, which improves access to high level medical care in underserved areas. The 23 chapters of this book present manifold examples of telemedicine treating both theoretical and practical foundations and application scenarios.

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